Case reports

Gallbladder agenesis diagnosed at laparoscopy

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Gallbladder agenesis is a rare but well recognized condition that was first described in 1701[1,2]. It is often asymptomatic and found at postmortem, however, it may be symptomatic and found at laparotomy. We report the first case found at laparoscopy for attempted laparoscopic cholecystectomy.

Case report

A 22 year old man had three attacks of right hypochondrial pain, nausea and vomiting, triggered by fatty foods. As a child he was diagnosed as having Gilbert's disease. Examination was unremarkable.

His serum bilirubin was 29 μmol/l (normal < 17 μmol/l). An abdominal ultrasound demonstrated a small contracted gallbladder containing a single calculus, together with a normal common bile duct (Figure 1). Therefore an elective laparoscopic cholecystectomy was scheduled. At laparoscopy, the gallbladder fossa was empty. The common bile was dissected from the liver to the duodenum but no gallbladder was identified. Operative cholangiography was attempted, but failed due to inability to safely secure the cannula in the common bile duct.

Postoperatively he made an uneventful recovery. A further abdominal ultrasound showed a normal common bile duct, but no gallbladder or calculi were seen. An ERCP demonstrated a short vestigial cystic duct stump, with otherwise normal extra and intrahepatic biliary anatomy (Figure 2). One year later he is well and asymptomatic.

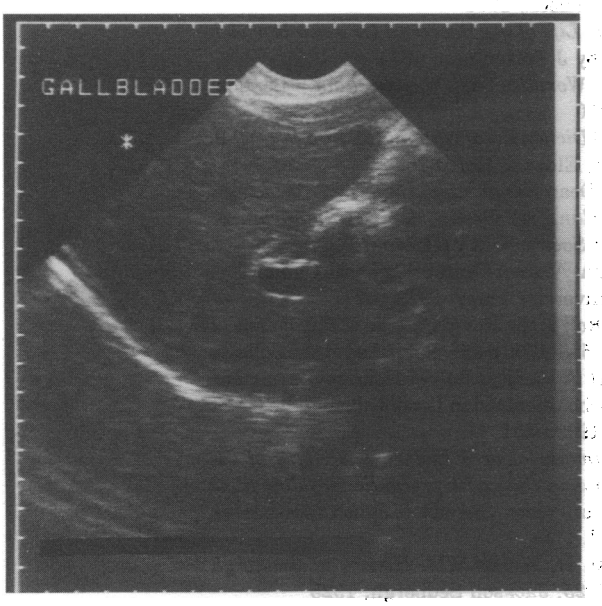

Figure 1. Preoperative abdominal ultrasound apparently demonstrating small contracted gallbladder with single calculus

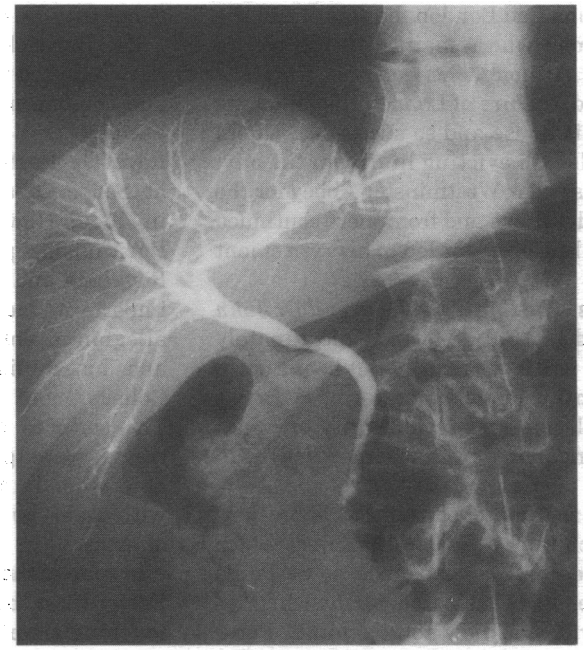

Figure 2. ERCP showing vestigial cystic duct with absent gallbladder

Discussion

The gallbladder develops from the caudal part of the hepatic diverticulum in the fourth week of intrauterine life[3]. Isolated gallbladder agenesis results when this cystic bud does not develop[3,4]. When cystic bud growth disrupts development between the sinus venosus cordis and the paired omphaloenteric and umbilical veins, gallbladder agenesis may occur together with cardiovascular and gastrointestinal abnormalities[5,6] and survival is rarely longer than 6 months[6]. Isolated gallbladder agenesis occurs in approximately 0.02% of live births and may be a non sex linked trait with variable penetrance[7]. The female : male incidence is equal in postmortem studies[8], but is 3 : 1 in clinical studies[6], probably because diagnosis usually occurs at laparotomy for cholelithiasis, which is more common in women.

A laparoscopic cholecystectomy was attempted on the basis of right hypochondrial pain and the ultrasound findings[9]. Although patients with gallbladder agenesis are usually asymptomatic, a significant number experience pain which is indistinguishable from cholelithiasis pain[5,10]. Possible causes of this pain includes biliary dyskinesis, adhesions, primary duct stones and non-biliary pathology. Interestingly, most cases of pain associated with biliary agenesis resolve after exploratory laparotomy and this may be due to lysis of periportal adhesions[10]. Currently our patient is asymptomatic one year postoperatively. False ultrasound reports are usual in gallbladder agenesis due to periportal tissue and subphrenic folds being interpreted as the gallbladder or calculi[10]. This makes preoperative diagnosis of gallbladder agenesis virtually impossible. When the gallbladder is not found at operation, it may be present in an abnormal situation[11]. Thus the whole extrahepatic biliary tree must be visualized and an operative cholangiogram performed if possible. In our case, ERCP showed a vestigial cystic duct and excluded an abnormally sited gallbladder.

Finally our patient had a slightly elevated serum bilirubin, which occurs in approximately 40% of patients with gallbladder agenesis[9], and a previous misdiagnosis of Gilbert's disease. Therefore, we recommend that congenital gallbladder

atresia be considered in the differential diagnosis of Gilbert's disease.

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Nasal obstruction due to enchondromas in Ollier's disease

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Ollier's disease is characterized by multiple enchondromas and areas of dysplastic cartilage in the tubular bones of the upper and lower limbs. It is an uncommon, nonfamilial dyschondroplasia. Classically the disorder affects the upper limbs bilaterally, but only one lower limb. Its aetiology remains unclear[1]. Involvement of the facial skeleton and skull in Ollier's disease is uncommon. A case of Ollier's disease with involvement of the nasal skeleton is reported.

Case report

An 18-year-old woman was referred with a 2-year history of progressive nasal obstruction and alteration in the shape of her nose. She had been known to suffer from Ollier's disease from infancy and had undergone numerous orthopaedic procedures.

Examination showed asymmetrical broadening of her nasal dorsum. Anterior rhinoscopy suggested the deformity was due to a mass expanding her nasal septum. The remainder of the examination of her head and neck region was normal.

CT scanning in coronal and axial planes demonstrated two discrete masses of soft tissue density expanding the nasal septum (Figure 1). Her skull base was not involved.

It was planned to excise these presumptive chondromas through a lateral rhinotomy approach. However initial exploration through a standard septoplasty approach gave adequate exposure for a resection of the tumours. Her postoperative recovery was unremarkable.

Histological examination of the resected tissue revealed highly cellular cartilage, with pleomorphic nuclei. Although these features could be consistent with the tumours being low-grade chondrosarcomas, because of her underlying Ollier's disease it was felt these tumours were simple enchondromas.

She has remained in good health since the procedure 4 years ago, with no tumour recurrence detected by annual clinical assessment and CT scanning.

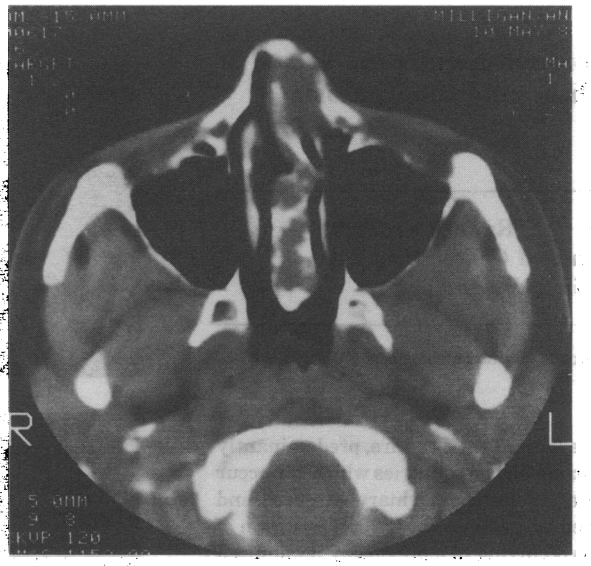

Figure 1. Axial CT scan showing expansion of the nasal septum

Discussion

Involvement of the facial skeleton is uncommon in Ollier's disease. This is probably because the majority of the bones of the skull and face ossify in membrane, not cartilage (the exceptions being the skull base and nasal skeleton). A literature search has revealed five reports of enchondromas or chondrosarcomas involving the skull base in patients with Ollier's disease[2-6]. Skull base tumours are more common in the related Maffucci's syndrome, where subcutaneous haemangiomas occur in association with dyschondroplasia[7]. There are no previous reports of tumours limited to tissues arising from the embryonic nasal capsule in either of these conditions. The involvement of the nasal septum alone in this case allowed an uncomplicated resection.

In common with the enchondromas which occur at other sites in Ollier's disease, the excised tissue in this case was highly cellular and exhibited nuclear pleomorphism. These features make it difficult to differentiate between benign enchondromas and low-grade chondrosarcomas. This problem is compounded by a tendency to malignant change in the multiple enchondromas of this disorder - the rates of malignant transformation quoted range from 30% to 50%[8,9].

In some series the malignant transformation rate may have been overstated, because of the atypical histological features of this condition. Pfleiderer *et al.* state the case they described, with a skull base tumour in a 20-year-old as the initial presentation of Ollier's disease, was probably misinterpreted as a low-grade chondrosarcoma until the